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Case Report

# A Trichoblastoma versus Basal Cell Carcinoma: A Case Report of a Basaloid Neoplasm in a Nevus Sebaceus of Jadassohn

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#### Abstract

Trichoblastoma is a rare benign biphasic, epithelial and mesenchymal, adnexal tumor recapitulating the hair bulb. It can be developed alone or rarely associated with nevus sebaceus of Jadassohn. Microscopically, it can mimic basal cell carcinoma. Awareness of their clinical, histological, and immunohistochemical characteristics is essential for proper pathological interpretation. This paper discusses a case of nevus sebaceus-associated trichoblatoma in a 13-year-old girl with histological features mimicking basal cell carcinoma.

Keywords: Nevus sebaceus of Jadassohn, organoid nevus, trichoblastoma, basal cell carcinoma.

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# **INTRODUCTION**

Nevus sebaceus of Jadassohn (NSJ) is a benign congenital hamartoma of the skin characterized by malformed epidermal, hair follicular, sebaceous, and other adnexal structures [1]. Benign and malignant cutaneous neoplasms have been reported with this skin disease, with trichoblastoma being the most frequently described neoplasm [2]. Trichoblastoma is a benign basaloid tumor with microscopic features recapitulating the hair follicular development. The microscopic features may overlap with basal cell carcinoma, a malignant basaloid tumor [3].

A case of trichoblastoma developed in nevus sebaceus of Jadassohn with emphasis on the clinical, histological, and immunohistochemical features is highlighted in this context.

#### **CASE REPORT**

A 13-year-old girl, not known to have other medical illnesses, presented with a hairless forehead yellow-colored skin lesion. This lesion was excised and

sent in a 10% neutral buffered formalin (NFB) container for histopathologic examination. The gross examination revealed a 3.5 x 0.7 cm skin ellipse containing a plaque-like irregular lesion with a yellow hue. Microscopic examination revealed a hyperkeratotic and papillomatous epidermis with morphologically abnormal dermal sebaceous glands that present high within the dermis and communicate directly with the epidermal surface. Also noted are markedly diminished hair follicles and characteristically heterotopic apocrine glands (figure 1). Incidentally, within serial sections, a focal dome-shaped basaloid lesion with peripheral epidermal collarette was found within multiple serial sections. It consists of sheets and cords of basaloid cells with peripheral palisading in a prominent wellcircumscribed bland cellular stromal component with papillary mesenchymal body formation. The diagnosis of trichoblastoma was supported histologically by the absence of stromal mucin and retraction artifacts and immunohistochemically by retained CK20 positive Merkel cells (figure 2).

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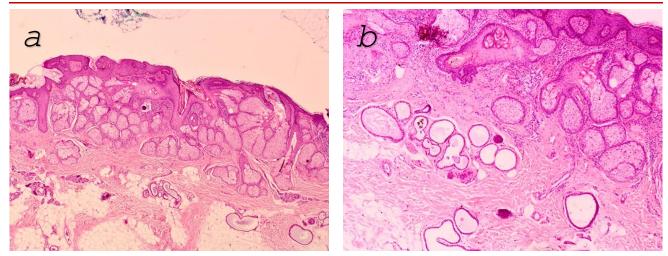


Fig-1: Microscopic pictures: showing hyperplastic sebaceous glands that present high in the dermis and open directly into the epidermal surface, H&E 100x (a), and scattered dermal heterotopic apocrine and dilated eccrine ducts, H&E 200x. Note the abnormally diminished hair follicles (a&b).

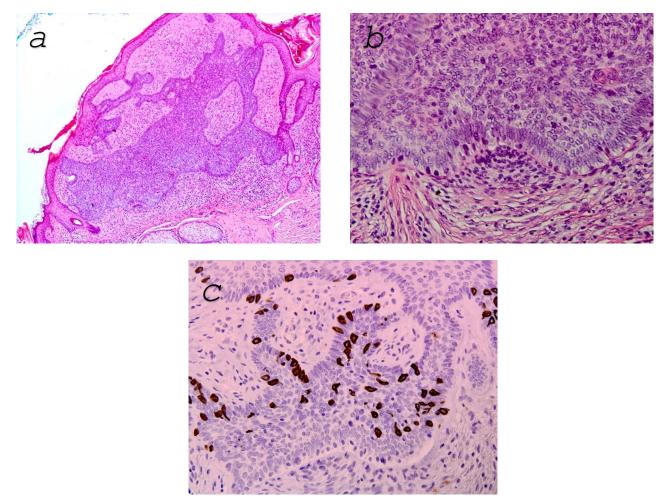


Fig-2: Microscopic pictures: Showing a basaloid tumor with peripheral nuclear palisading embedded in the cellular fibrotic stroma, H&E 100x and 400x, respectively (a&b). Note the characteristic stromal cells condensation that pushes into the basaloid nest, forming papillary mesenchymal body (b). CK20 positive Merkel cells are highlighted in a variable number throughout the lesion(c).

# DISCUSSION

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Nevus sebaceus of Jadassohn (NSJ), also known as "organoid nevus," has been recognized as an entity by Jadassohn, a dermatologist, since 1895, when he first described an early childhood forehead or scalp lesions that have a distinct waxy nodular surface [4]. It is a benign congenital hamartoma of the skin characterized by malformed adnexal structures, namely pilosebaceous units and apocrine glands. When patients grow up, these lesions become clinically apparent; moreover, at puberty, the sebaceous glands enlarge and become oily and vacuolated under the influence of gonadal androgens. The histopathological abnormalities are often present in the epidermis, hair follicles, sebaceous, and apocrine glands [5].

In a study by Kamyab-Hesari et al. [6], various histopathological features in 168 cases of NSJ were statistically analyzed, including epidermal, dermal, and adnexal abnormalities. Epidermal acanthosis and papillomatosis, followed basal by layer hyperpigmentation, spongiosis, and parakeratosis, were the most frequent epidermal changes. Chronic perivascular and perifollicular lymphocytic infiltration was commonly encountered in the lesional dermis. Sebaceous glands hyperplasia and immature hair follicles, followed by ectopic apocrine glands and eccrine ducts abnormalities, were the most encountered skin adnexal malformations. Nine of the patients in this (5.3%) developed secondary study neoplasms; trichoblastoma was the most prevalent (2.4%).

The incidence of secondary neoplasms in NSJ has been reported to be as high as 22.5% by Idriss and Elston [7], 14.4% by Cribier et al. [8], and 8.5% by Hsu et al. [9], respectively. These studies concluded that benign tumors are by far the most common secondary neoplasms. The most frequently encountered benign tumors were trichoblastoma (2-7%),papilleferum syringocystadenoma (3-5%),and trichilemmoma (1.6%). Secondary malignancies are rare, and to the best of my knowledge, only a few cases have been reported in the literature. Basal cell carcinoma was the most frequent secondary malignant neoplasm (0.8-1.1%) [7-9].

Trichoblastoma is a benign epithelialmesenchymal skin tumor with hair follicular differentiation. It was first described as an entity by Headington as a neoplasm that recapitulates the hair bulb's germ cells and dermal papillary mesenchyme [10]. Lesions usually present as an asymptomatic, solitary, well-circumscribed skin-colored nodule with a predilection for the head and neck area, most frequently scalp. Lesions are slow-growing and can be present for several years before being biopsied, and they can grow to be as large as 3 cm or even larger [11,12].

At the microscopic level, they are composed of dermal nests of uniform blue basaloid cells with

peripheral palisading encased by follicular derived dense cellular stroma that looks distinctive from the adjacent less cellular dermal connective tissue. They need to be differentiated from the more aggressive and infiltrative tumor, i.e., basal cell carcinoma, as these two tumors may show similar low power morphology. What is unique about trichoblastoma is the presence of the dense cellular stroma that tends to wrap and condensate right next to the basaloid nests, with characteristic stromal cells invaginations into the basaloid nests, forming papillary mesenchymal bodies [5,11]. Contrary to tricbhoblastoma, the stroma of basal cell carcinoma is usually less cellular and characterized by the presence of mucin-filled artifactual clefts [13].

The utilization of immunohistochemical stains can be helpful in complex cases. CK20 is a useful marker that can differentiate trichoblastoma from basal cell carcinoma. It can stain Merkel cells in the tumors follicular differentiation. Scattered with hair populations of Merkel cells (CK20 positive) can be detected in trichoblastoma rather in basal cell carcinoma (CK20 negative) [14]. Other markers that can be used for this purpose are BCL2, CD10, androgen receptors, and PHLDA1 [11]. BCL2 is only positive in the peripheral cells in trichoblastoma while showing diffuse positivity in basal cell carcinoma. CD10 only stains the stroma of the tricoblatoma while only showing an epithelial positivity in basal cell carcinoma [11,15]. Androgen staining is often negative in trichoblstoma and positive in basal cell carcinoma [11]. The follicular stem cell marker pleckstrin homologylike domain, family A, member 1 (PHLDA1), is a new helpful marker that differentiates trichoblastoma (positive) from BCC (negative) [16].

### **CONCLUSION**

Patients with NSJ are at risk of developing several benign and malignant tumors. Trichoblastoma is the most common neoplasm arising in NJS and is a rare benign skin tumor that needs to be differentiated from basal cell carcinoma. Awareness of these mimickers' clinical, histological, and immunohistochemical features is essential for proper histopathological interpretation.

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